

Dear Colleague:

This is the first publication of this newsletter. The purpose is to share information with physicians and other health care providers, medical records professionals, administrators/ policy makers, and all individuals in your community interested in birth defects in Kentucky.

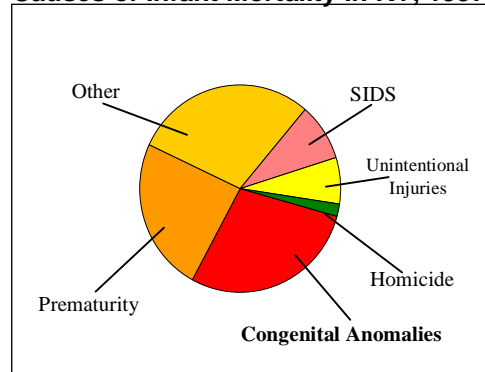
Congenital anomalies are the leading cause of death for Kentucky's children from birth to age one. There were 382 infant deaths in 1997 of which 27.5% were caused by birth defects. This fact places Kentucky as one of the top five states in birth defects-specific infant mortality. Also, the cost of disability due to loss of individual potential and medical expenses are tremendous.

The Kentucky Birth Surveillance Registry (KBSR) collects information on all children in Kentucky from birth to age five with congenital anomalies or disabling conditions. A surveillance system can only be effective with quality data. Therefore your role in paying special attention to reporting child case conditions defined by the Registry cannot be overestimated. The data collected by KBSR are used for determining the incidence of birth anomalies; enabling identification of potential etiology; planning interventions; and offering linkage of children and their families to services. Thus, it is critical that hospital medical records professionals, physicians, and other health care providers work together to assure accurate coding and reporting of birth defects.

You can help our efforts by noting cases that should be reported to the Registry and be being very specific when completing your patient hospital discharge face sheet. As needed, please call the KBSR, at (502) 564-2154 for further information. Thank you for your commitment to reducing the incidence of birth defects in Kentucky's children.

Steve Davis, M.D.
Director, Division of Adult and Child Health
Kentucky Department for Public Health

Causes of Infant Mortality in KY, 1997



Source: Kentucky Child Fatality Review

Did you know?



In the United States, birth defects have been the leading cause of infant mortality for 20 years



Birth defects account for more than 1 in 5 infant deaths in the US



86% of American women don't know that folic acid can help prevent certain birth defects

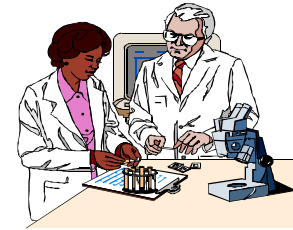


70% of American women don't get the recommended 0.4 mg of folic acid daily

In This Issue:

- Condition Highlight: Neural Tube Defects
- Genetics Corner: Why a Birth Defects Registry Program?
- Word from the March of Dimes
- KBSR Reportable Codes
- Birth Defects on the Internet
- Thalidomide: Back on the US Market
- Announcements

Genetics Corner



Why is an accurate birth defects registry important?

By: Bryan Hall, M.D.

A fictitious doctor at a state university medical school notes that the neonatal intensive care unit (NICU) has seen “a lot” of neonates with Gastroschisis, a serious congenital abdominal wall defect which allows the intestines to remain partially outside the abdomen. They have seen ten cases in the past year when the incidence of Gastroschisis in the U.S. is 1 in 10,000 liveborns. This means the NICU, which is a referral source for about half the state’s births (52,000 per year), would ordinarily expect 2.5 cases per year (25,000 births at 1 in 10,000 frequency = 2.5 cases). Should he call the state’s Vital Statistics Branch to see if they had any specific data for the state, or could he approach the Birth Defects Branch of the Centers for Disease Control in Atlanta and ask whether or not a cluster analysis should be done? Does the state have accurate data to make a comparison?

The above dilemma is a recurring theme because birth defects data reported to state vital statistics, on birth certificates, may be inaccurate with error rates of 30% - 50%. It is of critical importance to identify clusters and/or outbreaks of similar birth defects. Such defects may be the result of exposure to an environmental or drug teratogen, which could be eliminated, and the associated birth defect(s) prevented. Longitudinal data will allow for accurate comparisons when evaluating incidence and prevalence.

Additionally, the diagnostic code (ICD-9) for Gastroschisis is the same as for Omphalocele (756.79). A discharge diagnosis with the code 756.79 could represent either disorder. Chart reviews would be necessary to tell which of these two very different disorders each patient actually had.

The development of the Kentucky Birth Surveillance Registry (KBSR) offers the potential to deal with such problems by more

accurately documenting the incidence, prevalence and trends of birth defects over time. The incoming data will be more detailed and verifiable as they are received by the KBSR. Consequently, clusters or outbreaks of birth defects can be recognized and analyzed earlier, making the decision to investigate further more secure.

ICD-9 diagnostic codes are not always specific enough to adequately identify the condition. ICD-9 codes are the basis of ascertainment of birth defects reported to KBSR. The KBSR plans to use abstractors with knowledge of genetics to review medical records to clarify birth defect diagnoses. Other informational sources such as screening programs for metabolic diseases and hearing loss, and the genetic services of the University of Kentucky and University of Louisville can be linked to the KBSR to achieve maximum ascertainment and accuracy.

The KBSR has been a long time in coming. A lot of hard work by dedicated people has brought it fruition. The opportunity to achieve accurate documentation that can be used for prevention, treatment, early intervention, and state planning for Kentucky’s children with birth defects, is now here. State and federal resources will get us to the next level.

Dr. Hall is a Dysmorphologist at the University of Kentucky Medical Center and the Medical Director for the KBSR.



Condition Highlight

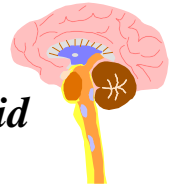
By: Lorraine Keil, M.S.N., R.N.

Neural Tube Defects (NTDs) affect approximately 4,000 pregnancies in the United States each year, with an annual cost in Kentucky of 12 million dollars. NTDs occur during the first 28 days of pregnancy, often before a woman knows she is pregnant.

NTDs are characterized by a failure of the skin around the spine to close. Children with this condition have lifelong physical disabilities with a loss of motor movement.

The most revolutionary discovery that has occurred over the last 20 years in the field of birth defects, is the role that folic acid plays in the prevention of this defect. Studies have demonstrated that 50% - 70% of Spina Bifida can be prevented. Based on these findings, the United States Public Health Service published

Preventing Neural Tube Defects through Folic Acid Supplementation



recommendations in the Fall of 1992 that all women of reproductive age take 0.4 mg of folic acid daily. This is the amount contained in most multi-vitamins. In March 1996 the Food and Drug Administration (FDA) agreed to fortify any grain labeled “enriched” with folic acid.

Despite these efforts, when the Kentucky Behavioral Risk Survey was conducted in 1996, only 37% of the respondents knew folic acid could help prevent birth defects. This revealed the need for more vigorous educational efforts. NTD surveillance and folic acid education programs must continue to promote folic acid. The KBSR will be crucial in measuring the success of these programs.

Lorraine Keil is the Director of Nursing for the Commission for children with Special Health Care Needs.

Neural Tube Defects in Kentucky

The incidence of neural tube defects (NTDs) in Kentucky is estimated to be 1 per 500 births compared with the national rate of 1 per 1,500 births. The lifetime cost per child with spina bifida is estimated to be \$294,000.

Why does Kentucky have such a high incidence of NTDs?

- ♣ English, Scottish, Irish and Welsh heritages are associated with higher incidences of NTDs as is Hispanic heritage. Kentucky has a high population of people with these European backgrounds and in recent years has seen a sharp increase in its Hispanic population.
- ♣ Low socioeconomic level is associated with the risk factor of poor nutrition and lack of prenatal care.
- ♣ Kentucky's consanguinity rate is 7% compared to 5% for the rest of the country.
- ♣ In 1996, eight (8) Kentucky children died from anencephaly.

The Kentucky Birth Surveillance Registry

By: Linda Lancaster, R.N., M.P.A.

The Kentucky Birth Surveillance Registry (KBSR) was developed through collaboration between various hospitals, the Kentucky Hospital Association, KBSR Advisory Council, service and advocacy organizations and the Department for Public Health to develop an information collection method. The KBSR collects information from acute care and birthing hospitals through one central data clearinghouse on all children from birth to five years of age who are diagnosed with any structural, functional, or biochemical abnormality determined genetically or induced during gestation.

On April 21, 1998, President Clinton signed into law the Birth Defects Prevention Act of 1998 (P.L. 105-168). This act signifies national concern for children and families and the potentially devastating effects of birth defects. Although the Centers for Disease Control (CDC) has supported birth defects surveillance for many years, this Act establishes formal authority within the CDC to provide surveillance, research and services aimed at the prevention of birth defects.

The KBSR operates under the authority of KRS 211.651-670 with statewide data collection for surveillance beginning in April of 1996. The Kentucky statute and regulations require appropriate and accurate hospital reporting of information leading to the identification of children with birth defects.

Information gained through surveillance assists the State and communities in planning and policy development in an effort to address priority health needs and gaps in service for these children with birth defects and their families. The KBSR plays an essential role in informing and educating communities and families in Kentucky about health behaviors that can reduce the risk of having a baby with a birth defect.

KBSR data will help determine the number of children born in Kentucky with certain birth defects. Additionally, KBSR aggregate data may help in the study of contributing factors and possible causes of particular defects. Data can be analyzed by Kentucky region to identify

(continued on page 5, column 1)

KBSR ICD-9 Condition Categories (ICD-9 CM Codes)

All Congenital Anomalies	740 –759		
<i>Examples:</i>			
Microcephaly	742.1	Umbilical Hernia	553.1
Macrocephaly	742.4	Teratogen (noxious influences)	760.7
Upper GI Anomalies	750	Fetal Alcohol Syndrome	760.71
Lower GI Anomalies	756.7	Narcotics	760.72
		Hallucinogenic Agents	760.73
Metabolic/Storage Disorders	270-279*	Cocaine	760.75
*Excluding codes 274, 276, 278		Small for Gestational Age	764
		Hemolytic Disease of Newborn	773
Dwarfism, Not Elsewhere Classified	259.4	Infant of Diabetic Mother	775
Hereditary Hemolytic Anemia	282	Seizures	779*
Neurologic Disorders of Brain and Cord	334	*To allow for coding error	780.3
Cerebral Palsy	343	Failure to Thrive	783.4

A complete listing of ICD-9CM codes may be obtained by contacting KBSR administration at (502)564-2154 or E-mail KBSR@mail.state.ky.us.

The Kentucky Birth Surveillance Registry

(continued from page 4, column 2)

possible areas where a particular birth defect incidence is high or increased. If a certain region is noted to have a high rate, further investigation can be initiated through the registry.

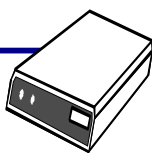
Specific information as to type of birth defect is identified through ICD-9 CM diagnostic codes from hospital admissions and outpatient services. This information is provided by the hospital and directed through a state data clearinghouse which formats the data for Registry

use. These data are then matched with birth and death certificates to collect additional information. Confidentiality measures are applied throughout this process.

The KBSR is beginning a new phase of implementation. Having recently received federal funding for program enhancement, the KBSR is positioned to provide not only detailed reports, but also to be a resource for families to learn about services which can allow their child to meet his/her greatest development potential.

Linda Lancaster is the program director for the Kentucky Birth Surveillance Registry.

UB-92 Reporting



Submissions to the KBSR **MUST INCLUDE** the following items in addition to the items requested by the Health Policy and Analysis Branch:

- First and Last Name of the the Child (Patient)
- Complete Address of the Mother/Parent
- First and Last Name of the Insured
- Social Security Number of the Insured

Incomplete submissions will be returned to the facility with a request to resubmit these claims with the required information.

A Word from the March of Dimes

By: Katrina Adams

About 150,000 babies are born with a birth defect each year in the United States (about 4% of live births). In an average week in Kentucky 1,007 babies are born. Eight of those babies will die before their first birthday. Presently, the infant mortality rate stands at 7.2 per 1,000 live births (1997). The goal for the year 2000 is 7.0. Birth defects have been the leading cause of infant mortality in the United States for the past 20 years, accounting for more than 1 in 5 infant deaths.

The March of Dimes has dedicated 10 million dollars to a national folic acid campaign to reduce the number of babies born with neural tube defects (NTDs). By making all women aware of the benefit of folic acid, they expect to reduce the number of NTDs by 30% by the year 2001.

Katrina Adams is the Director of Program Services for the March of Dimes, Kentucky East Chapter.

Folic Acid Campaign Manual

A resource manual to help in the development and implementation of folic acid promotion/NTD prevention campaigns is now available from the Centers for Disease Control and Prevention.

The manual, Preventing Neural Tube Defects: A Prevention Model and Resource Guide for folic acid campaigns is available by e-mail at: flo@cdc.gov or phone 770-488-7190.

*"Flo" - The Folic
Acid Awareness
Campaign Logo*

News and Announcements

Thalidomide: Approved On U.S. Market

Thalidomide was widely prescribed in Europe in the 1950s to treat anxiety, insomnia and, in pregnant women, morning sickness. It was marketed in numerous countries including Japan, Australia, and Canada. Thalidomide was withdrawn from the market in the early 1960's when doctors learned that it **caused devastating birth defects**.

In 1998 the FDA gave approval for thalidomide to be marketed for the treatment of erythema nodosum leprosum (ENL), a complication of leprosy, a disfiguring skin disorder involving loss of feeling that can lead to paralysis. The brand name drug, Thalomid, is being tested for a range of other diseases (studies with Thalomid in the areas of cancer, HIV/AIDS, dermatology and gastroenterology are now being supported).

According to a recent survey, two-thirds of people of childbearing age are not familiar with thalidomide. Because thalidomide may become more available to women capable of having children, they need to be warned about the serious risks of birth defects.

Due to thalidomide's danger to a developing baby, the FDA requires special safeguards never before imposed with any drug. To prevent fetal exposure to thalidomide, the drug's manufacturer has developed the System for Thalidomide Education and Prescribing Safety (STEPS) program. Only physicians who are registered with the STEPS program may prescribe thalidomide and those patients - both male and female - must comply with mandatory contraceptive measures, patient registration and patient surveys. Thalidomide may be dispensed only by licensed pharmacists who are registered in the S.T.E.P.S. program and have been educated to understand the risk of severe birth defects if thalidomide is used during pregnancy.

In addition, female patients' prescriptions will not be filled without a physician's written

report of a negative pregnancy test that has been conducted within 24 hours of starting thalidomide therapy. Pregnancy testing will continue to be required weekly during the first month of use, then monthly thereafter in women with regular cycles, or every two weeks if cycles are irregular. Prescriptions are only for one month's supply.

All patients must participate in a mandatory registry that will provide follow-up to detect any adverse effects of using thalidomide and will hopefully identify areas in which safeguards need to be improved, if problems occur.

This article was extracted from the March of Dimes homepage, www.modimes.org.

Kentucky Children Health Insurance Program (KCHIP)

Beginning July 1, 1998, a special program called KCHIP (Kentucky Children Health Insurance Program) will provide low income Kentucky children health insurance coverage. This program is designed to ensure that all children have access to adequate health care coverage. Proper care will help assure our children's healthy growth and development. KCHIP is for families with incomes up to 150 percent of the federal poverty level. Any child, birth through eighteen, without health insurance may be eligible. Services available include physician, hospital, well child preventive including immunization and prenatal care, behavioral health, prescriptions, vision care, dental, home health and many others. Services in the community will be provided by physicians and other professionals and agencies through a Managed Care Organization. For more information about KCHIP call the Kentucky Department for Medicaid Services at (800) 635-2570.

Folic Acid Awareness Campaign

The National Folic Acid Campaign is a collaborative effort of organizations that belong to the National Council on Folic Acid, a coalition led by the March of Dimes.

The mission of the Council is to reduce birth defects by promoting the use of folic acid. In addition to increasing the daily consumption of folic acid by women of childbearing age, the campaign also aims to:

- make folic acid awareness a routine and standard part of the delivery of preventive health care services to women, and
- increase the level and availability of folic acid in food.

Each state will develop its own folic acid campaign with guidance from the National Council on Folic Acid, The March of Dimes and the Centers for Disease Control.

The Division of Adult and Child Health, through the leadership of Dr. Steve Davis is very committed to the Kentucky Campaign.

Watch for Kentucky's Folic Acid Campaign Kickoff in September 1999!



Birth Defects on the Internet

State Birth Defects Registries:

Arizona:

(www.hs.state.az.us/edc/abdmp/Abdmp.htm)

This site contains case definitions, descriptions of the data sources and procedures, as well as data on selected defects for the state by race and county.

Colorado:

(www.cdphe.state.co.us/dc/dccrcsn.html)

Contains a program description, general state and county level data tables, and information on how to request data or other reports.

Hawaii:

(members.aol.com/entropynot/hbdp.html)

The site contains a description of the program including background information, case definitions, activities, collaborative studies and publications.

Iowa: (www.pme.huiowa.edu/birth.html)

This site provides background information and a program description. There are several pages discussing health and environmental issues as well as a page with brief descriptions of current research projects.

New Jersey:

(www.state.nj.us/health/fhs/scregis.htm)

This site provides a brief description of the the Registry, the reporting laws and a detailed description of reporting requirements.

Birth Defects Related Sites:

March of Dimes: (www.modimes.org)

This site provides information on the organization along with descriptions of its community services, education programs, birth defects information, and infant health statistics. There is also the latest issue of its monthly statistical newsletter.

Medical Genetics, University of Kansas Medical Center:

(www.kumc.edu/gec/support/groups/html)

This site provides information on genetic conditions and birth defects, along with links to national and international organizations, genetic counselors and clinical geneticists.



Seventh Annual Infant/Toddler Conference

July 28-30

Executive Inn West, Louisville, KY

For more information:

Germaine O'Connell or Linda Wainscott

DMHMRS First Steps

100 Fair Oaks Lane, 4E-E

Frankfort, KY 40621

(502) 564-7722

42nd Annual Maternal and Child Health Conference

September 8-10

Galt House, Louisville, KY

For more information:

KY Department for Public Health

Division of Adult and Child Health

275 E. Main Street, HS 2GW-A

Frankfort, KY 40621-0001

(502) 564-2154

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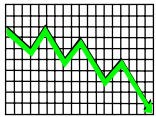


To be added to the mailing list or to request more information on KBSR, please contact us at (502) 564-2154 or KBSR@mail.state.ky.us.

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**Kentucky
Birth
Surveillance
Registry**

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